seem to be essential to the disease. In his own cases referred to, the thyroid had been enlarged. This was undoubtedly a remarkable case, analogous to those of the enlargement of lateral half of the body. Of this he had seen two cases. If these cases were myœdematous we must revise our notions of that affection.

DR. PUTNAM considered that transition cases were always especially valuable. Pathologically, the affection described would appear to be related to myxœdema, but clinically the relation did not appear distinct.

DR. DERCUM stated that the fluid which exuded in his case resembled lymph rather than the denser fluid of myxœdema. He thought that the case was allied to the type without being a true myxœdema. He referred to the fact, however, that in quite a number of cases of myxœdema, mental symptoms had been marked. In some cases too, the thyroid had not been enlarged but wanting. He thought it difficult to determine minor degrees of enlargement of thyroid through the skin.

The following paper was then read :

SUBACUTE PROGRESSIVE POLYMYOSITIS.

BY GEO. W. JACOBY, M.D.
OF NEW YORK.

The increase in our knowledge of the various affections of the muscles has, as is well known, during the last ten years assumed proportions which could hardly have been foreseen. That certain muscular affections are due to disease of the anterior columns of the spinal cord, that others are due to disease of the peripheral nerves, and that still others are entirely independent of such affections, but are primarily localized in the muscles themselves, are facts which have assumed the character of axioms. All the attention necessary for the attainment of these results has, however, been given to the chronic forms of muscular disorders, the forms generally described under the designation of degenerative atrophy, and the acute and subacute disorders, particularly the inflammatory ones, those showing the characteristics of inflammation as seen in other organs, have been markedly neglected. It is therefore our purpose by means of this paper to call to your attention the little that has been done in this direction, and by the report of a remarkable case, one presenting *intra vitam* all the typical signs of inflammation,invading successively nearly all the muscles of the body, ending fatally, and showing microscopically not only the characteristics of acute and subacute inflammation, but also many of those found in the chronic forms of primary dystrophies, to endeavor to trace a relationship, perhaps a connection between the acute and chronic primary muscular affections. The case is as follows:

Patient, F. H., male, æt. 35. Family history is unimportant. Married; three children, one dead. His wife has had no miscarriages. Neither he nor his wife have had syphilis, nor do they show any suspicious marks. Habits as regards alcohol and tobacco good. The patient was always always perfectly healthy until about four years ago. At that time he had an attack of intermittent fever, for which and during subsequent illnesses he was treated by Dr. H. J. Boldt, of this city, by whom the patient was kindly referred to us. Four or five months later he had an attack of pleurisy, and subsequently he was prostrated with a severe attack of typhoid fever which incapacitated him for about three months. The recovery from the typhoid was complete, and he remained perfectly well for more than a year. During this time he attended to his work, that of a machinist, without the loss of a single day, and felt as well as in his healthiest days, weighing more than he did prior to his illness. After the lapse of this year (two and a half years ago), having been in good health the day previously, he complained of a feeling of tension in the right calf. The gastrocnemious muscle "seemed to be severely inflamed." The skin was red and shining and there existed pain to superficial as well as to deep pressure. Slight œdema was also present. The diagnosis of a cellulitis was made and a deep incision effected under antiseptic precautions. No pus was present. The wound healed by first intention, and the patient seemed improved. After a few weeks a similar condition occurred in the left calf, and the right one also returned to the state which existed prior to the incision. The tension increased in both legs and was particularly noticeable in the morning. Finally, on account of this tension and swelling of the muscles, he found difficulty in bending the legs at the knees. The condition varied from day to day, but if one leg seemed somewhat better, the other one was worse. Then the thigh of the right side became similarly affected, and this was followed by the same condition in the left thigh. Then the flexor surfaces of both forearms became involved, and finally the biceps of both arms.

I saw the patient for the first time on May 11, 1888, when he presented the following *status*: He is a tall, thin cachetic-looking individual. The facial muscles are normal. Eyes and pupils, tongue and speech normal. The deltoid and trapezius on the left side were enlarged and painful to pressure. So also the biceps of the left arm and all the muscles on the flexor surface of the forearm. The right arm is similarly affected but to a slight degree. The entire left arm feels harder than the right. The skin over both forearms on the flexor surface shows a slight erythematous blush, which is also more marked upon the left than upon the right side. On the hands the balls of both thumbs seem somewhat tender to pressure, but not swollen.

The lower extremities are both equally affected. The quadriceps cruris and the adductors are very much enlarged and painful to pressure, as are also the gastrocnemii. The muscles of the feet are not affected. The motions of the patient are impeded in accordance with the muscles involved. He can execute all movements, but those requiring action of the affected muscles show a certain effort which at once attracts attention. The affected muscles have a peculiar feeling, recalling very much, as do also all movements of the lower extremities, cases of pseudo-hypertrophic paralysis. They however feel more elastic. The tendons of all the involved muscles can be plainly felt as tense cords. Slight œdema of the skin, which however does not take the impression of the finger is noticeable over all the affected muscles. The pain on pressure is confined entirely to the body of the muscle. Pressure over the nerve trunks does not cause any pain.

Electrical examinations showed the excitability of the affected muscles to be greatly reduced to both currents. A current of 15 m. a. was requisite for the production of cathodic closure contraction. No reaction of degeneration in any of the muscles. Slight patellar tendon reflexes obtainable with great difficulty. Sensation normal. No bladder affection. The mechanical excitability of the affected muscles was not increased, perhaps reduced. Fibrillary twitchings were not present. There was no atrophy discernable in any parts of the body. Heart, lungs and abdominal organs normal. Measurements of the legs and thighs gave the following results:

Thigh, left, upper part, - -	58	cm.
" " middle, - -	53	"
" " lowers, - -	43	"
Leg left, below the knee, -	35	"
" " middle of calf, - -	41	"

The right leg and thigh gave slightly smaller figures.

On the 16th of May, Dr. Kammerer, of this city, kindly excised for me two pieces of muscle, one piece from the gastrocnemius of the right leg and the other piece from the supinator longus of the left forearm. Both wounds healed by first intention.

During the following fortnight there was no particular change in his condition. One day he would be able to walk around, and another day he would be confined to his bed. Then his condition grew decidedly worse, the swelling of the extremities increased, the muscles became exquisitely painful to the touch, but the joints remained freely moveable and free from exudation.

On July 3d, Dr. Boldt, requesting me to see the patient again, writes:

"He has acute cirrhosis of the liver. The stage of enlargement has passed and the organ for the past ten days is getting smaller again. The connective tissue seems to be rapidly shrinking. Examinations of the urine only show decrease of urinary salts."

July 4th.—I found the condition as follows:

Abdomen tense with ascitic fluid. The face and body very much emaciated. Sweats profusely. The affected muscles in consequence of the general emaciation appear much more swollen than actual measurements prove them to be. The figures obtained by measurement are essentially the same as those above given, with the exception of the lower parts of the thigh. There is very great pain upon motion and to pressure. The attachment ends of the muscles are approximated and the tendons stand out hard and unyielding. The skin has a deep erysipelatous appearance over all the implicated muscles. Minute capillary hæmorrhages are also seen along the borders of these discolored territories. In addition to the muscles previously mentioned as being affected, the glutei, the muscles of the back, the intercostals and the external obliques of the abdomen are also more or less involved. That, however, which at once attracts attention on account of the contrast produced is the atrophy which has invaded certain muscles and which is assuredly not due to the general emaciation. Upon both thighs it is seen that the lower part of the quadriceps, particularly the vasti are very much wasted. This is particularly marked upon the left side, which was previously the larger of the two. The contrast caused by the wasting of the vasti and the great hypertrophy of the rectus, abductors and glutei was very marked. A similar contrast was also observable in the arms where the biceps and triceps were very much enlarged while the deltoid and anterior part of the trapezius, which had also been affected were, particularly upon the left side, almost entirely atrophied. The shoulder joints were movable. The dorsal interossei of both hands and the thenar muscles also atrophied, together with the ball of the thumb of the left hand, that of the right hand being swollen and painful.

July 18th.—Condition the same. Consolidation of apex of left lung. No other muscles involved by the atrophy.

July 25th.—General condition much worse. Has difficulty in swallowing. Tongue painful and swollen. Cannot protrude it beyond the edge of the teeth.

Aug. 2d.—Pain in swallowing so great that only by the greatest efforts can he be induced to take a little milk. His condition is a truly deplorable one. Active movements of any kind are impossible. The head cannot be lifted from the pillow and even a slight rotation of the head causes great distress. The forearms are flexed almost at right angles to the arms. Pressure-force of hands *nil.* The legs flexed upon the thighs and cannot be straightened. The feet themselves are freely movable. Extended passive movements cannot be made on account of the resistance of the tense tendons. Face and eyes not affected. Sensation not affected. Pupils, sight, smell and hearing normal. Sensorium perfectly clear.

Aug. 12th.—Death due to general marasmus, and disorders of respiration.

Owing to my absence from town at the time of death, a post-mortem was unfortunately not obtained.

The pieces of muscle exsected from the leg and arm were immediately placed in a half per cent. solution of chromic acid. When sufficiently hardened they were embedded in celloidin and cross and longitudinal sections made. These sections were then stained with an ammoniacal carmine solution and mounted in glycerine. The histological changes found will now be described.

In transverse sections of the piece of muscle exsected from the leg (Fig. I.), the small size of the single muscle fibres, without any noticable decrease in their number in each individual bundle, is marked. I do not, however, attribute this feature to the morbid process itself, since with the exception of a moderate increase of the perimysium internum, the fibres have retained their mutual flattening, and even the sarcolemma sheath appears to be preserved, except in a few muscle fibres which are highly altered. In addition we also, with low powers, at once notice a distinct broadening of the perimysium externum as also of the perimysium internum, the latter not being so conspicuous and not so uniformly observable as the former.

The newly formed connective tissue which goes to make up the augmented perimysium externum, is either myxomatous or fibrous in structure. The latter again is either loose and delicate or dense and fibrous, being then made up of comparatively coarse bundles. In both varieties of the perimysium externum we frequently meet with the following two features.

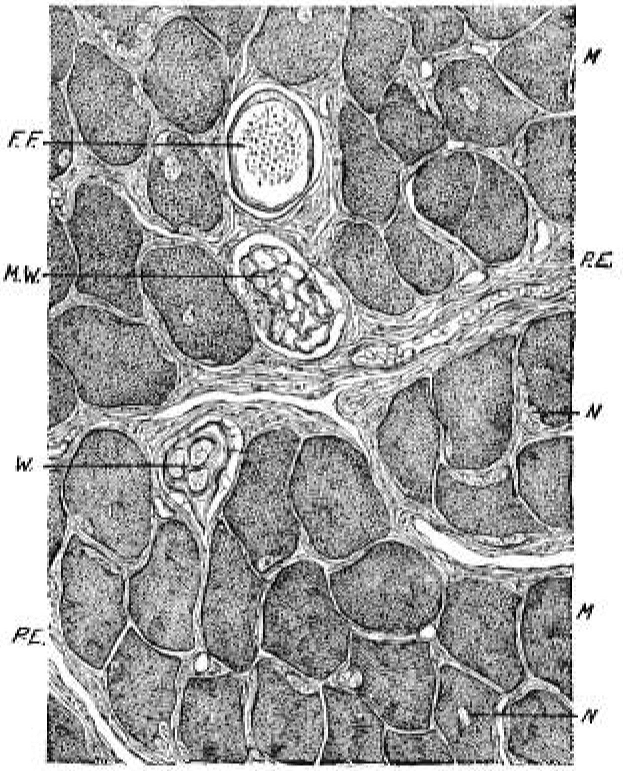

Fig. I.—Transverse Section x 200.

M. M. Muscle fibres of average size in partly transverse, partly slightly oblique sections.
P. E. P. E. Perimysium externum with numerous capillaries.
N. N. Nuclei, some in centre, some at periphery of muscle.
W. Muscle fibre in waxy degeneration, holding three protoplasmic bodies.
M. W. Cluster of medullary corpuscles in waxy degeneration.
F. F. Muscle fibre in waxy. possibly combined with fatty degeneration.

Firstly, fat globules in a more or less regular arrangement; and, secondly, waxy degeneration of the bundles of fibrous connective tissue, in some places very pronounced. The perimysium internum, although as already mentioned, augmented, is made up entirely of a delicate fibrous connective tissue, carrying numerous blood-vessels, but nowhere exhibiting fat globules or waxy degeneration.

The muscle fibres themselves apart, from their small size, show peculiar changes, which never invade all the fibres of a single bundle, but only a limited number, which varies from one to eight.

These affected fibres are either transformed into an apparently homogeneous mass, with traces of nuclei in it and still surrounded by an unchanged sarcolemma sheath, or the muscle is transformed into a highly refracting so-called waxy mass which shows distinct nuclei, or is entirely homogeneous, or homogeneous only at the periphery and granular in the centre. It is not uncommon to see muscle fibres which are partly normal in appearance and partly waxy or homogeneous. Many of the fibres contain nuclei in their interior, which, when fallen out, leave open spaces or vacuoles. The number of such nuclei and vacuoles varies from one to half a dozen or more in a single fibre. Other fibres again are in part or entirely broken up into smaller lumps with or without nuclei (Fig. II.). The staining of the specimens with the ammoniacal carmine solution also yielded peculiar results, inasmuch as many of the fibres appear deeply stained in one part and unstained or yellow (owing to the chromic acid solution) in another. Such variations in color occur even in distinctly waxy fibres, some of which are deeply carminized, others again appearing unstained. Since hæmorrhage had taken place at the moment of excision of the muscle and a subsequent saturation with the blood plasma must have occurred, the differences in the reaction to this staining reagent would, it appears to us, in our case be deserving of but little attention.

From the appearances thus far described, as seen with low powers, it follows that a plastic or formative inflammation has invaded the muscle, being most conspicuous through the augmentation of the interstitial connective tissue, more especially of that of the perimysium externum. This process is known under the term of hyperplastic perimyostitis, a process very prominent in our case of pseudohypertrophy from which microscopical studies were presented to this Association last year.

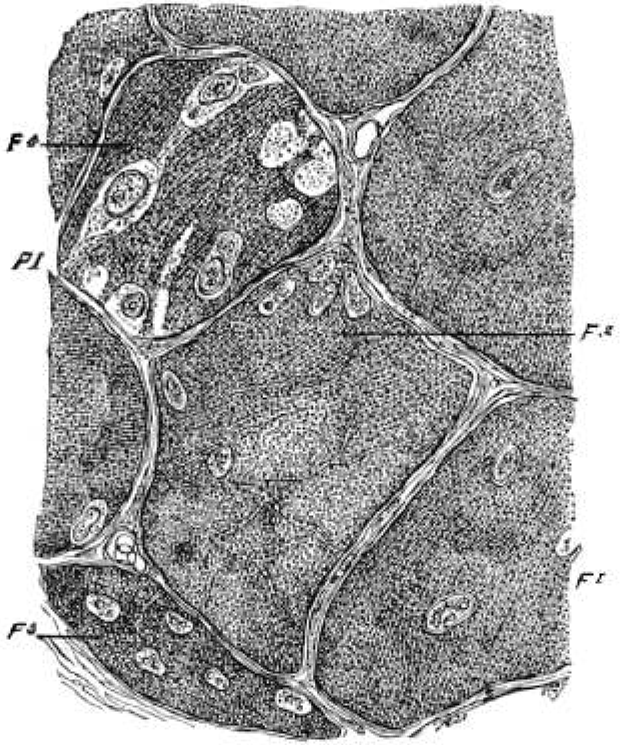

FIG. II.—Transverse Section x 800.

F.1. Muscle fibre with central nuclei.

F 2. Muscle fibre with augmented peripheral nuclei, showing indication of breaking up into muscle plates.

F,3. Muscle fibre, holding five central nuclei, each one corresponding to a muscle plate, the whole resembling a myeloplax. The sarcous elements enlarged and crowded.

F.4. Muscle fibre transformed to a great extent into partly nucleated, partly non-nucleated protoplasm.

PI. Perimysium internum almost unchanged.

At the same time marked changes must have occurred in the muscle fibres themselves, leading to a breaking up of the individual fibres, their contractile matter into lumps, and terminating in waxy degeneration—a process which is recognized as a parenchymatous one since Virchow, and meriting the name of "myositis."

The question now arises, What relation in our case does the myositis bear to the perimyositis? In other words, how much of the process is interstitial and how much parenchymatous?

This question can easily be settled by resorting to higher powers (500 diameters). The first feature which impresses the observer is the augmentation of the nuclei. In the normal muscle, nuclei are seen at the periphery of the muscle fibre close beneath the sarcolemma, but not in large numbers. In our case, muscle fibres are seen holding sharply defined nuclei scattered throughout the body of the fibre without regularity, and surrounded by a light rim, possibly due to shrinkage; whereas the contractile matter is apparently little changed and still exhibits the granulation produced by the sarcous elements.

As central nuclei, in the fully developed fibre, are known to occur in human beings only in the muscle of the heart, the presence of a larger number of nuclei within the fibre, as such, would indicate a pathological process. When the nuclei are much increased in number, the transverse section of the muscle fibre may assume the aspect of a so-called giant cell, whereby the regular arrangement of the sarcous elements is lost and the granulation has become irregular. In this case the sarcolemma sheath is still present and still allows the boundary of each single muscle fibre to be easily defined. In the next stage the muscle fibre breaks up into a number of indifferent or medullary corpuscles, many of which still contain nuclei, either singly or in groups. The boundary of a muscle fibre has in this stage to a great extent lost its sharpness; and since the adjacent perimysium is largely composed of similar bodies not markedly nucleated, no strict boundary line can be drawn between the muscle and the perimysium. The inference, however, almost forces itself upon us, that the original muscle tissue, after having split up into medullary corpuscles, loses its specific structure and becomes transformed into fibrous connective tissue, adding, as it were, a certain amount to the hyperplastic perimysium itself. This inference impresses itself upon us all the more strongly when we see fields of fibrous perimysium with traces of previous muscle fibres composed of indifferent corpuscles, and finally fields of considerable extent between slightly changed muscle fibres, embracing fibres in waxy degeneration.

Still more instructive are for our purpose muscle fibres which in part are unchanged, in part, on the contrary, are transformed into a coarsely granular mass, which has entirely lost the regular arrangement of the sarcous elements, and is provided with a varying number of nuclei. The presence of the sarcolemma is a positive proof that the morbid changes have taken place in a portion of the contractile substance of a single muscle fibre only. Such a partial transformation is by no means a rare occurrence.

The appearance of a number of nuclei within the muscle fibre is not the only way in which the morbid changes are inaugurated. Sometimes a muscle fibre breaks up partly or *in toto* into clusters and lumps, apparently due to an increase of the contractile matter and its grouping into homogenous masses. Such lumps may likewise grow up to the size of nucleated medullary corpuscles, indicative perhaps of a more acute course of the process. This process we must define as a myositis, a process leading first to a reduction of the muscle fibre into its embryonal constituents, the so-called muscle plates or sarcoplasts. One of the terminations of myositis has already been described, namely, transformation of the contractile matter into fibrous connective tissue, or perimysium. In addition to this termination we find two others present in our case, namely, fatty and waxy degeneration of the muscle fibres.

Here and there transverse sections of muscle fibres are met with, which contain vacuoles in their centres, varying from one to five in number, and enclosed by a sharply defined capsule. Such vacuoles differ in appearance from empty spaces by dropped-out nuclei. Although the fat is mostly extracted from the specimen, owing to the use of alcohol and ether for the purpose of embedding, the comparison with analogous formations in the perimysium admits the diagnosis of the formation of fat in the middle of the muscle fibre. It is also this treatment with ether and alcohol which renders it impossible to say whether or not fibres lacking the carmine stain, being yellow, highly refractive, and split up into clusters by irregular crevices, are in fatty degeneration. We know, however, that the sarcous elements may be directly transformed into fat granules, by the coalescence of which, appearances are produced identical with those seen in these specimens.

The second and more common termination of the myositis process is waxy degeneration. In almost every bundle we find one or more muscle fibres transformed into a highly refractive mass, known under the name of "waxy" or "amylaceous." The manner in which this metamorphosis is produced can easily be traced. At first we notice a breaking up of the muscle fibre into a number of medullary corpuscles, whereby the sarcolemma sheath remains unaltered. Such corpuscles are saturated with or infiltrated by a peculiarly changed blood serum, which renders them refractive, glossy, and homogeneous. Often a number of medullary corpuscles coalesce into a lumpy, homogeneous mass, in which faint traces of the boundaries of the previous medullary corpuscles are discernible. In the highest degree of waxy degeneration we see the homogeneous mass almost continuous, holding in its interior star-shaped or oblong protoplasmic bodies not in waxy degeneration, so that the appearance of cartilaginous tissue is brought forth. If the number of protoplasmic bodies prevails over the waxy basis substance, the latter may produce a framework similar to a honeycomb, the sarcolemma under these conditions being entirely lost.

Longitudinal sections of the muscle of the leg show all the changes described above as present in transverse sections (Figs. III. and IV.). Especially conspicuous is the small size of all fibres, these at the same time being split up lengthwise into minute fibrillæ, and only exceptionally exhibiting a distinct transverse striation. The size of the sarcous elements is strikingly small. The augmentation of the number of the nuclei is obviously less noticeable in the longitudinal than in the transverse sections, but here and there chains or rows of nuclei are seen.

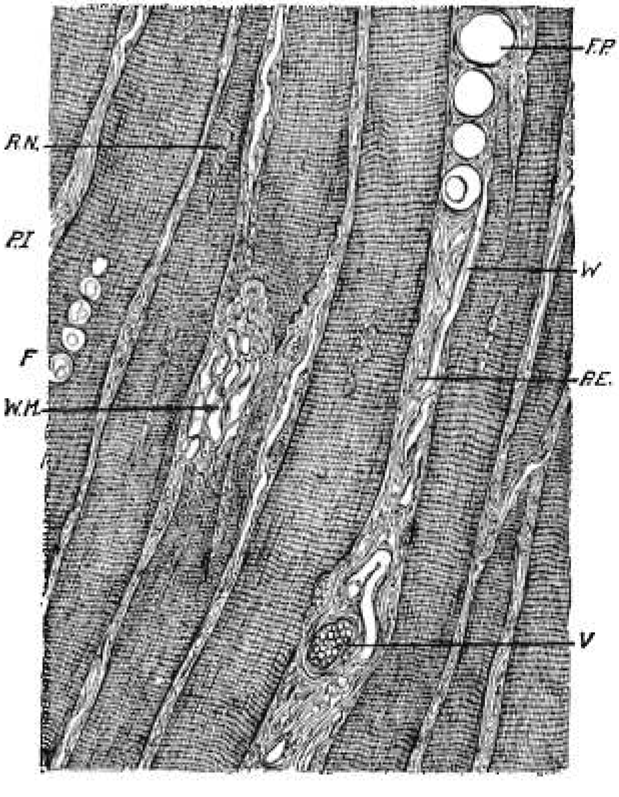

FIG. III.—Longitudinal Section x 200.

Pe. Perimysium externum, considerably augmented; composed of coarse bundles of fibrous connective tissue.

V. Vein, engorged with blood.

F. P. Fat globules in external perimysium.

P. I. Perimysium internum, slightly augmented.

R. N. Rows of nuclei.

F. Row of nuclei.

F. Row of **fat** globules ? in centre of muscle fibre.

W. M. Clusters of medullary corpuscles in waxy degeneration In the vicinity of this cluster, the gradual transformation of the muscle tissue into inflammatory corpuscles and the destruction consequent destruction of the muscle tissue is marked.

W. Peripheral portion of the muscle fibre in marked waxy degeneration.

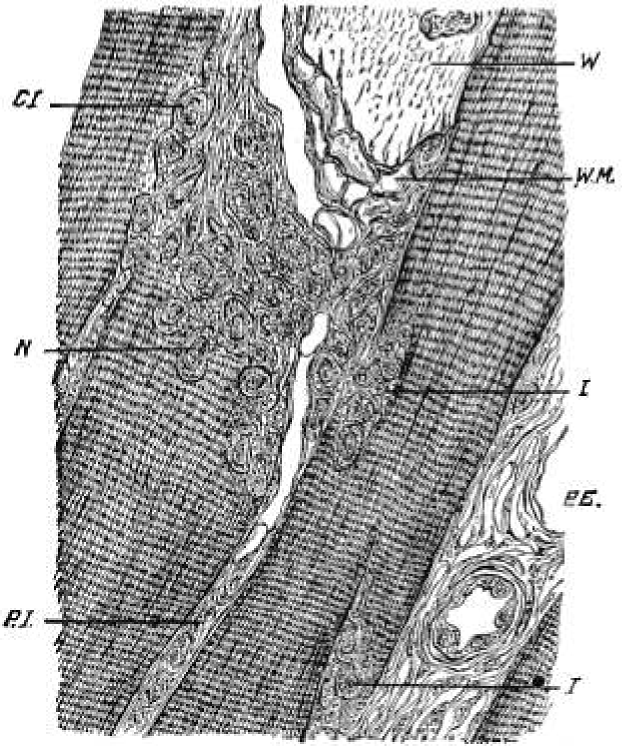

FIG. IV.—Longitudinal Section x 800.

P. E. Perimysium externum, broadened, composed of coarse bundles of fibrous connective tissue freely vascularized.

P. I. Perimysium internum, transformed into inflammatory or medullary corpuscles.

I. I. Groups of inflammatory corpuscles, obviously arisen from previous contractile tissue

N. Nucleated inflammatory corpuscles imbedded in muscle tissue.

C. I. Clusters of inflammatory corpuscles in bay like excavations of the contractile tissue, in part spindle shaped, in transition to fibrous connective tissue. The medullary tissue transversed by a large, probably newly formed capillary blood-vessel.

Both the perimysium internum and externum have increased in size, especially the externum. That this increase is not a mere augmentation of the fibrous connective tissue, but is the result of an active participation of the contractile tissue, can plainly be demonstrated in longitudinal sections. Here we not infrequently encounter muscle fibres whose body terminates almost abruptly, the fibrillar structure being lost and replaced by medullary tissue. It also happens that only a limited portion of the muscle fibre appears transformed into rows of medullary corpuscles, blending with the contractile substance proper. This contractile substance first shows a breaking up into the muscle plates, and subsequently into rows or clusters of medullary corpuscles. Such clusters are seen to appear in the middle of a muscle fibre, being then bordered by an almost unchanged muscle tissue. The medullary tissue assists in the new formation of both external and internal perimysium, whereby the bulk of the muscle fibre is diminished and that of the fibrous connective tissue augmented.

Waxy degeneration is a marked feature both in the perimysium and in the muscle fibres. In the former, even the middle and adventitial coats of the arteries and the adventitial coats of the veins are subject to waxy degeneration. In no place of the muscle tissue itself has this degeneration attacked the muscle fibre in toto. Lumps showing the characteristic high refraction, as a rule are seen only in the axes of muscle fibres, as though representing prolongations of them. This fact, more particularly however the appearances seen in transverse section of the muscle, forces upon us the conviction that it is the contractile tissue itself which has become waxy, after having first been broken up into medullary tissue.

Numerous medullated nerve fibres are visible in longitudinal as well as in transverse sections. These are seen singly as also in bundles. Many of these nerves appear to be unchanged, whereas in others an increase of the nuclei of Schwann's sheath can be seen, and in still others the perineurium internum appears to be more or less crowded with medullary or inflammatory corpuscles, features which we are accustomed to see in neuritis and perineuritis.

How many medullated nerve fibres have been destroyed by being transformed into fibrous connective tissue, we are unable to say. The large number of unchanged nerve fibres, however, strongly points towards the view that the process of neuritis and perineuritis is not the primary one, but altogether secondary to the myositis and perimyositis. In the same manner as a certain amount of contractile tissue is transformed into fibrous connective tissue, with the intervening stage of inflammatory or medullary tissue, so also the nerves are at least in part changed, first to medullary, and finally to fibrous connective tissue.

The muscle of the arm shows changes identical with those observed in that of the leg, the only difference between the two being that the process is less extensive and less intensive in the former than in the latter.

In summing up the results of this examination, I would condense them as follows :

1. The process is an acute myositis and perimyositis, grafted on a chronic plastic or formative process.
2. The plastic process has led to a new formation of connective tissue of the perimysium externum as well as of the perimysium internum. This process became progressive by repeated recurrences of myositis, leading to a diminution of the contractile, and to an increase of the fibrous connective, tissues.
3. The result of the chronic process is fatty and waxy degeneration. Both have invaded the perimysium, as well as the muscle tissue proper.
4. Neuritis and perineuritis are secondary to myositis and perimyositis.

Before entering into consideration of the case itself, if we now take a brief review of that which has heretofore been done in acute disorders of the muscular tissue, we will see that, notwithstanding inflammation of muscles ought *a priori* to occur as easily as inflammation of other tissues, as a matter of fact, it was universally supposed that such inflammation of the muscles themselves was impossible, until Virchow,[1] in his remarkable thesis of 1852, showed this view to be erroneous. Even at present there are many observers to whom the existence of a primary myositis savors somewhat of a myth. This condition of affairs can only be due to the fact that, although inflammation of the muscles, be it due to operative measures or traumatism of various kinds, or to propagation of the inflammatory process from neighboring parts, is often produced, nevertheless the myositis itself rarely comes nnder the observation of the physician. The slight forms cause so little inconvenience to the patients, and the severe forms are of so rare occurrence, that even when these severe forms are encountered *intra vitam*, a disunity of opinion as regards the diagnosis and character of the affection has been the result. Our knowledge of the microscopical changes produced by the process has, with perhaps the exception of the more minute changes, remained the same as it was left by Virchow. According to this observer, the pathological changes occur either in the muscular fibre itself, in the interstitial connective tissue, or in both, and it is often impossible to determine from which of these the process primarily originated. The contractile substance becomes involved in a number of regressive changes—hyperæmia, turbid swelling, change in color, loss of longitudinal and transverse striation, then fatty or waxy degeneration, or, in very acute cases, destruction of the fibrillæ and softening.

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[1] *Virchow*, Ueber Parenchymatoser Entzundung, Archiv. f. Patholog. Anat. und Physiologie, 1882, Vol. 4, p. 261-323.

The processes occurring in the interstitial connective tissue are of a more active nature; here we have the formation of pus, granulation and fat tissue, or tendon and bone tissue, which substances were supposed to force themselves between or supplant the contractile elements. Thus, according to the tendency of the changes of the medullary corpuscles, we have either a fibrous, a purulent, or an ossifying myositis. The category in which our case belongs, microscopically, is, as already stated, that of a parenchymatous myositis.

Upon entering upon the clinical aspect of this case we are confronted by an important question. Is the affection here a primary or a secondary one? Of secondary affections of the muscles not due to local causes, we have pain and swelling due to muscular rheumatism. Etiologically and anatomically, we know nothing as to the character of this affection. Clinically, we only know that beyond the pain and swelling there is no analogy between the two affections.

So also after polyarticular rheumatism occasionally, many muscles are affected in a similar manner; but in our case there is no history of joint involvement at any time.

After variola, Wagner[1] has seen similar occurrences.

The muscular implications occurring in consequence of scorbutus, glanders and syphilis need only be mentioned to be disposed of. Not so is it with the muscular changes occurring during typhoid fever. To these more than a passing notice must be given. Changes in the muscles of typhoid fever patients, resembling very much the changes described by Virchow as above stated, were observed by Zenker[2] in 1864. He found degeneration of the striated fibres in the most varied groups of muscles, and as has been shown by later observers these changes are almost as characteristic as the changes of the intestinal mucous membranes. The adductors of the thighs and the recti abdominis are the muscles chiefly involved. A medium or pronounced degree of waxy degeneration is here found, sometimes associated with granular degeneration. A certain number of fibres still retain their normal transverse striation. The extraordinary fragility of the muscles is shown by the frequency with which transverse ruptures are encountered. Microscopically, Zenker differentiates the granular and waxy degeneration, the first produced by interspersion of finest molecules in the contractile muscular substance, the latter consisting of a transformation of the contractile substance into a waxy, homogeneous, colorless, shining mass, with complete disappearance of the transverse striæ and destruction of the muscular nuclei, the sarcolemma alone remaining intact. In addition Zenker also noted degeneration in the form of discoid separation and fibrillary splitting. The connective tissue, vessels and nerves show no changes, but in many cases a proliferation of nuclei takes place in the perimysium. Although the comparison of the microscopical findings in our case with those of Zenker show many points of identity, still it is evident that there are also points of difference. But even were the results of the microscopical examination absolutely identical, we would not be justified in considering the processes as clinically analogous. Indeed we have long believed and have publicly expressed the opinion that from an examination of muscles alone, it is impossible to decide to which clinical variety of muscular disorder a given case belongs.[1] This idea also receives support from Waldeyer,[2] who in myositis, mechanico-experimentally produced, found the same histological changes as in typhoid myositis. Therefore it is necessary in a review of conditions analogous to those presented in our case also to give particular attention to the clinical aspects.

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[1] Archiv. der Heilkunde, 1872, p. 107, Vol. xiii.

[2] *Zenker*, Ueber der Veranderungen der Wilkurlichen Muskeln in Typhus Abdominalis, Leipzig, 1864.

The clinical symptoms differ materially in our case and in typhoid myositis. In the latter the course is of a light character with slight pains and slight tenderness to pressure, whereas in our case the other extremes were noted. We have no hesitancy in excluding the fact of our patient having had typhoid fever over a year prior to the occurrence of the described affection, as entirely free from any etiological significance.

The next class of cases which bear a certain analogy to ours are those described by Scriba.[3] The principal point emphasized by these cases is that we may have an acute myositis due to infectious inferences. Here the infection was caused in three cases by a furuncle and in one by a carious tooth. The fifth case is valueless for comparison, as it did not present a pure myositic picture. These cases presented slight fever, severe pain in the muscles affected, with swelling. The affected muscles were greatly increased in size and were of a bony consistency to the touch, and could be traced from origin to insertion, like a plaster cast. The process was always limited to isolated muscles and the contiguous muscles were soft and painless. The cases all ended in complete cure in several weeks, either with or without suppuration. In the second case microscopical examination of a teased preparation of a piece of muscle was made. The transverse striation was found to be almost entirely absent. The sarcolemma were from two to three times larger than is normally the case. The perimysium externum was slightly infiltrated with medullary corpuscles and the perimysium internum was only very slightly implicated. Scriba considers the affection very rare even in Japan, and was unable to obtain the history of a single similar case from other physicians. In Europe, also, the affection was entirely unknown prior to this publication. The points of variance between these cases and ours are also evident.

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[1] Discussion, N. Y. Neurological Society, March 5, 1888.

[2] *Waldeyer*, Ueber die Veranderungen der Quergestreiften Muskeln bei der Entzundung und dem Typhus Process, etc., Virchow's Archiv., Vol. 34.

[3] *Scriba, J.*, Myositis Acuta, Deutsche Zeitschrift f. Chirurgie, Leipzig, 1885, p. 497–502.

This completes the review of the acute secondary inflammatory muscular disorders. Of the acute primary or so-called idiopathic disorders still less is known.

Under the name of acute and subacute myositis, a class of cases has been described by various observers, Strauss,[1] Clark,[2] Treves,[3] Devis,[4] Walther,[5] Hepp[6] and others, giving as causes cold, blows and muscular over-exertion.

Nosologically this affection stands in the same rank as acute periostitis and osteomyelitis, with which it has in many cases been confounded. It is undoubtedly an acute infectious disease with a tendency to suppuration differently localized according to the preceding infection. All the muscles of the body become affected. A typical course cannot be given, but it is undoubtedly a serious disease, beginning with dull, increasing pains, which render the use of the affected muscle impossible. The muscle swells, becomes hard, abnormal positions ensue (due to approximation of the attachments), suppuration takes place, causing single large or several small abscesses, and the severe cases end fatally, the lighter ones recovering.

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[1] *Strauss*, Jaccond's Dictionnaire, Vol. xxiii., p. 362.

[2] *Clark, F. W.*, British Med. Journal, 1887, ii., p. 69.

[3] *Treves*, Brit. Med. Journ., 1886, ii., p. 1215.

[4] *Devis*, Tr. Pathol. Soc. London, 1881, p. 273–75.

[5] *Walther*, Deutsche Zeitsch. f. Chirurgie, 1885, p. 285–87. Gives table of 19 cases, of which cases 12 (*Maas*) 15, 16, 17 and 18 (Scriba) do not belong here, but must be classed as secondary affections.

[6] *Hepp, Paul*, Ueber einen Fall von acuter Parenchymatoser Myositis, welche Geschwulste bildete und Fluctuation vortauschte, Berliner Klinische Wochenschrift, 1887, p. 389.

Only one more class of cases remains to be described, and to this class, of which only three cases have been thus far reported, our case belongs. As these cases are of such rare occurrence, and they all have been described within the last two years, I may be pardoned if I enter upon them somewhat closely. A few cases have been described, prior to these, nnder other names, which probably also belong to this class.

These cases clearly show the existence of an acute inflammation, rapidly affecting nearly all of the voluntary muscles, characterized chiefly by pain and swelling, and ending in death within a more or less short period of time. They show us that we may have an acute primary progressive inflammation of the muscles, the chronic form of which alone has been hitherto known. The first case is that of Wagner.[1] Female, æt. 34. Years ago had typhoid. Now tuberculous. Since end of June, without exposure to draught or cold, pains in joints, back, neck, shoulders, and legs. Received in hospital, July 19th, 1886. Well nourished, strongly built. Muscles of neck painful to pressure. Stiffness in both shoulder joints, but freedom from pain. Slight œdema of back of hands and forearms as well.

July 24th.—Paræsthesia in both forearms. Œdema the same. Nerves not sensitive to pressure.

July 28th.—Arms also swollen, forearms most. Skin tense, so that contours of muscles could not be distinguished. Muscles feel doughy. Movements impeded.

August 7th.—Swelling has decreased, Erysipelatous redness on flexor surface of left arm.

August 17th.—Redness has spread.

August 24th.—The thighs have become affected. Cough and dyspnœa.

August 29th.—Disorders of deglutition and spontaneous attacks of dyspnœa. Death.

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[1] *Wagner, E.*, Ein Fall von acuter Polymyositis, Deutsches Archiv. f. Klin. Medizin, Vol. 40, 1887, p. 241.

Autopsy.—Extensive muscular disease. Slight œdema of both upper extremities. Slight tuberculous phthisis of lungs. Diffuse bronchitis. Severe emphysema. Hyperæmia of brain and cord. The muscles which were microscopically changed were the biceps, triceps, and brachialis internus. These were of a peculiar, stiff consistency, but at the same time more fragile than normally. In the forearm the flexors are chiefly affected, but irregularly, spots and stripes of very pale appearance alternating with darker parts. Nearly all the muscles except the glutei, muscles of the calf, and the abdominal muscles, are affected. Brain, spinal cord, and peripheral nerves normal. The majority of specimens for microscopical examination were taken from the biceps. Serous infiltration, fatty and waxy degeneration of the muscle fibres, were the characteristics most marked. The interstitial connective tissue showed proliferation of nuclei, and single multinucleated giant cells were also observed. Processes of new formation were found in the muscular substance, as well as in the perimysium. Only in a few places was the perimysium internum increased. The adventitia of the small arteries was somewhat rich in nuclei. The nerve endings in the muscles were not examined.

The second case is that of Unverricht.[1] The patient was a male, æt. 24, who had been well until two weeks prior to presentation. Etiologically nothing. For a few days dragging pains in arms and legs, which increased in severity until walking was impossible.

Examination showed a medium-sized, well nourished individual, of good muscular development. Organs of chest and abdomen normal. Complains of pains in all extremities and back. Motion of extremities intact, but painful; force normal, no paræsthesia. No fever, nor disorders of digestion. After a week, slight swellings of the extremities and also of the face. Increase of the swelling. Muscles painful to pressure. The joints free from exudation, and painless. At this stage the extremities were swollen to thick, formless masses, whereas the body was not affected. Later, the thorax became painful to pressure, and swollen. Three weeks after entrance to hospital, pains in swallowing; fluid food alone could be taken. Thoracic muscles very painful, breathing abdominal, pneumonia, death.

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[1] *Unverricht*, Polymyositis acuta progressive, Zeitschrift f. Klinische Med., 1887, p. 534.

Autopsy showed brain and medulla normal. The muscles showed extensive and remarkable changes, chiefly limited to certain groups. Color in general, dark reddish brown, but upon section many muscles are found colored in irregular spots. Pale, somewhat transparent, light gray strips alternate with dark red spots. All these muscles are swollen and fragile. The changes are most marked in the extensors of the extremities, whereas the flexors are almost entirely unaffected. The pectoralis major affected in parts. The deltoids extensively, biceps slightly. Extensors of forearms very much, triceps free, muscles of ball of thumb free. Extensors of thighs and toes very much, but flexors of thighs and legs only slightly affected. Abdominal muscles, muscles of back, and intercostals slightly implicated. Muscles of tongue, eyes, and diaphragm not affected. The idea of trichinasis had been expressed, but not a single trichina was found.

Microscopically, the results corresponded to the macroscopical examination. The interstitial connective tissue was infiltrated with medullary bodies. Muscular substance in all stages of degeneration, some places showing normal muscle, and the immediately contiguous place being severely degenerated. Loss of transverse striation; hygroscopic; granular and waxy degeneration. In other places fatty degeneration was found. Regeneration, and increase of muscular nuclei not present. Spinal cord and peripheral nerves were unchanged.

The third case is that of Hepp.[1] Patient, female, æt. 36. Family history good. Previous history unimportant. Sick since March, 1886; did not feel well during two to three weeks; then eruption on face of red, thickly settled, elevated and non-itching spots. Next day spread to back, neck, and mucous membrane of mouth. After a week, disappeared. Then increasing pains in neck and back; legs swollen, but not the thighs. Took to bed. After eight days, swelling of face and right arm, then, after three days, of left arm. Swelling of face disappeared, that of arms increased. Spontaneous pains rare, but very severe upon slightest motion. Then stiffness of extremities; could not bend or extend arms; legs not so bad.

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[1] *Hepp, Paul,* Ueber pseudo Trichinose, eine besondere Form von acuter parenchymatoeser Polymyositis, Berl. Klin. Wochenschrift, 1887, p 297.

Examination shows slight redness of face and chest. Œdema of face, extremities, and chest; feet, ankles, and hands free. Arms swollen to a hard cylindrical mass; pectoralis major in contracture; hard, stiff, and non-elastic. Muscles of neck and lower extremities the same, but not so marked. No pains when quiet. Joints free; passive movements free and painless; extended movements difficult on account of contracture of muscles. Active movements not possible. Facial muscles and tongue not affected. Voice, pupils, sensorium, special senses not affected. Tendon reflexes absent. Affected muscles do not react to either electrical current. Increase of disorder, affection of muscles of deglutition, death on May 22d. Anatomically, affected muscles yellowish and pale. Everything else normal.

Microscopical Examination.—Absence of trichinæ. Waxy or hyaline degeneration near apparently normal fibres; others in which transverse striation indistinct, and in still others entirely absent. Others showed all stages of hyaline transformation, from the cylindrical hyaline fibre to complete disintegration into granular detritis. In no place granular or fatty degeneration. Spinal cord normal.

The following two cases have been published under other names, but probably also belong in this category. First, by Wagner.[1] Woman, æt. 43. Well until a few days prior to entrance into hospital.

Status, Oct. 17th, 1861.—Badly nourished, pale, small. Both arms very much swollen up to elbows. Forearms to hands less. Reddened, œdematous ; great pain on pressure. Lower extremities not swollen, but have a feeling of tension. Difficulty in swallowing fluids.

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[1] *Wagner, E.*, Fall einer seltenen Muskelkrankheit, Archiv. der Heilkunde, 1863, Vol. iv., 282 and 83. Additional remarks in the article on Polymyositis by the same author. Loc. Cit.

19th.—Swelling of upper extremities less.

20th.—More swelling.

21st.—Arms very much swollen. Death occurred during the following night.

Microscopical examination showed intense and extensive changes in all the muscles of the upper part of the body. Partly fatty metamorphosis, partly colloid degeneration. In many places both forms of degeneration were found in one and the same fibrillæ. Numerous diffuse, very small foci of pus corpuscles, and increase of the nuclei. The intermuscular and subcutaneous connective tissue did not show any proliferation of connective tissue bodies or pus.

Second Case.—Marchand.[1] Patient male, æt. 23. Sick since five weeks. Began with gradually increasing muscular pains and corresponding disorder of function. At the same time œdematous swelling without albuminuria occurred. Diagnosis of trichinosis. The respiratory muscles became affected and death ensued. The autopsy revealed an intensely diseased condition of the muscular system, not equably distributed, but affecting principally the extensors of the extremities, partly also the muscles of the chest and neck as well as the adductors and flexors of the thighs. The muscles were increased in size. No trichinæ were found. Microscopical examination showed granular and fatty as well as hyaline degeneration of the primitive fibres. Spinal cord and peripheral nerves normal.

Now, having found the proper place for our case, there are two questions, applicable to this entire class, which must be answered before any final conclusion as regards the position if any which it bears to the chronic forms of primary dystrophies can be arrived at. The one is the question of trichinosis in the production of analogous conditions, and the other is that of the relation which multiple neuritis bears to these cases. In all the cases above cited as being cases of polymyositis, the similarity of the symptoms to those observed in cases of trichinosis is marked, and all of the authors with the exception of Wagner, in his first case (1861), had even more than a suspicion that trichinæ were the cause of the severe muscular disorder. Also in two cases reported by Kussmaul and Maier,[1] which although accepted by others as cases of polymyositis, I have purposely excluded, this conviction is expressed. Hepp even goes so far as to give the name "pseudo-trichinosis" to the complaint, a name which has received its well-merited criticism and censure from others. In none of the cases, however, were trichinæ found in the muscles, except in Wagner's second case, and here in about three hundred muscular sections three trichinæ were found; these were encapsulated and estimated to be about four months old, so that also in this case their influence upon the production of the myositis must be excluded.

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[1] Breslauer Aerztliche Zeitschrift, 1880, p. 264.

In our case the question is effectually disposed of, not only by the absence of trichinæ from the microscopical specimens, but also on account of the long duration of the disease.

That multiple neuritis may produce symptoms similar to those observed in our case cannot be gainsaid. But if we take into consideration the facts, that during the entire course of the disease there was freedom from spontaneous pains; there was no pain produced by pressure over the nerve trunks; there were no disturbances of sensation; there was no actual paralysis of any muscles, the inhibition of movements being due entirely to the pain produced by them; there was no reaction of degeneration in any of the muscles, and, finally, considering the intense swelling of the muscles, we believe that neuritis as the primary affection may be unhesitatingly excluded. All of these negative symptoms taken together are valuable, a single one by itself of course proves nothing.

Owing to the changes found in a few of the intramuscular nerves it is to be greatly regretted that pieces of the larger nerves could not be obtained for examination, although, as above expressed, it is evident from the microscopical examination that the neuritis is secondary to the myositis.

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[1] Ueber eine bisher nicht beschriebene Erkrankung (Periarteritis nodosa) die mit morbus Brightu und rapid fortschreteinde algemeiner Muskellahmung einhergeht, Deutsches Archiv. f. Klin. Med., Vol. i., p. 484.

The frequently cited cases of combined neuritis and myositis described by Eisenlohr,[1] as well as the recent article of Senator[2] upon the same subject, go far to prove that the inflammatory process, whether it primarially originates in the nerves or in the muscles, does in many cases not remain confined to the primarily affected tissues, but secondarily involves the muscles if the nerves, the nerves if the muscles were originally involved. In Eisenlohr's and Senator's cases the presence of disorders of sensation with paralysis of the muscles, in addition to the other symptoms, make the primary involvement of the peripheral nerves probable.

Another question, and one which cannot be disposed of so easily, is the possible relationship between our case and cases like ours, and primary progressive muscular atrophy ; in short, is our case perhaps a case of progressive muscular atrophy in which the inflammatory stage is more marked, clinically, than usual, due to the myositis being more of a parenchymatous character ? Would we not possibly, if we examined our cases of the chronic forms with a view particularly to the history of the initial stage, find analogous conditions to those presented by our case, or at least a history of muscular rheumatism so-called ? In other words, are not some cases of primary progressive muscular atrophy the resultants of light cases of polymyositis parenchymatosis ? We believe that there is considerable evidence in favor of these views, and we do not stand entirely alone in possessing this opinion. Wagner insists and Unverricht admits the possibility of their cases being acute cases of primary progressive muscular atrophy. Cases, in fact, of the acutest possible kind, Wagner's cases showing a duration of three and eight weeks respectively, and Unverricht's a duration of six weeks.

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[1] *Eisenlohr*, Idiopathische subacute muskellahmung und atrophie, Centralblatt f. Nervenheilkunde, 1879, p. 100, Vol. ii.

[2] *Senator*, Acute multiple myositis bei neuritis, Deutsche Med. Wochenschrift, 1888, p. 449. Discussion in Verein f. Innere Medizin, meeting May 28, 1888. Berl. Klin. Wochenschrift, p. 493, 1888.

None of these cases however, showed any atrophy, but it cannot be said that this might not have developed if the life of the patients had been more prolonged. At any rate the proof of a relationship between these very acute cases and the chronic ones, would be a very difficut one to furnish, unless an intermediary can be found which will serve as a connecting link. This link we believe to possess in our case.

We admit that if we compare this case with typical ones of primary progressive muscular atrophy, the disparity is evident, but we must not forget that we are dealing with an unusual case, the assignment of which to any of the well known types of disease meets with difficulty. But let us compare the two affections, and the following points of similarity and dissimilarity will become apparent. The first objectionable point in our case is the late occurrence, namely, at the age of 32. Still, we know that rare cases of the pseudo-hypertrophic form do not begin until the eighteenth or twentieth year, and that Schultze's,[1] which, although a very unusual case, he classes under this form, did not begin until the twenty-seventh year, we also know that in the primary atrophic form the occurrence may take place as late as the sixtieth year.[2] We here speak of the pseudo-hypertrophic and atrophic forms, almost in one breath, because we all know that the separation of the two forms is not absolutely certain on account of intermediary cases, which cannot easily be placed in one or the other category.

The second objectionable point is the mode of origin. This in all cases of polymyositis, as also in ours, was acute, a mode decidedly at variance with that of progressive muscular atrophy. But even here, exceptions to this rule are furnished by the so-called rheumatic forms, a number of cases of which have been compiled by. Fredreich,[1] who has also added one of his own.

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[1] *Schultze, F.*, Ueber den mit Hypertrophie Verbundenen progressiven Muskelschwund, Wiesbaden, 1886.

[2] *Gowers*, Manual of Disorders of the Nervous System, 1888, p. 395.

The duration of the affection in the cases of Wagner, Unverricht and Hepp, was so short, that the main stumbling block for comparison seems to lie there. In one case the duration of two and a-half years is also short, but still it is a step towards the shortest duration of authentic cases of progressive muscular atrophy (8-10 years). As worthy of note is the fact that in these short cases death has generally been due to involvement of the respiratory muscles, as it was also the cause in ours.

Now the points of complete similarity may be mentioned.

(1.) There was in our case, the same symmetrical distribution of affected muscles as is found in cases of primary progressive muscular atrophy.

(2.) The degeneration always affected single muscles and parts of muscles (degeneration individuelle), and not entire muscular masses (atrophie en masse). This was very characteristic microscopically ; here this *dégeneration individuelle* could be traced down to the single bundles, where single fibres in each bundle were always seen to be affected, and not the entire bundle.

(3.) Sensory disorders were absent.

(4.) Reaction of degeneration was not present.

An endeavor to class the case among any of the so-called "types," taking into consideration the distribution of the atrophy, would be entirely fruitless, as the patient died so soon after the onset of the atrophy that the possible ultimate distribution of the same cannot even be surmised. Furthermore, we believe that the lines separating the various known groups are entirely too tightly drawn, which is clearly shown by such cases as Barsickow[2] and Zimmerlin.[3]

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[1] *Freidreich, N.*, Ueber Progressive Muskelatrophie, Berlin, 1873.

[2] *Barsicow*, Zwei Familien mit Lipomatosis muscular, progress. Diss. Halle, 1872. In one of these families the affected patients generally showed involvement of the mescles of the legs and back. but in one case the muscles of the *arms* and *back* were affected. In the other family the legs alone were usually affected, but in some members of the same family the shoulder muscles became affected first and then the muscles of the legs became involved. How are these cases to be classified ?

[3] *Zimmerlin*, Mendel's Centralblatt, 1885, No. 3. In one family, two cases of juvenile form and third beginning in face, with pseudo-hypertrophy of lower limbs.

Microscopically the results of the process in our case was seen to be the same as those in progressive muscular atrophy, namely, destruction of the muscle by inflammation as well as by degenerative processes. While last year, in our paper on pseudo-hypertrophy, we took occasion to express surprise at the complete similitude between the results of the microscopical examination in that case and those obtained by Friedreich[1] in a case of spinal progressive muscular atrophy, we this year must call attention to the great analogy presented microscopically by our case of a year ago, with the one of to-day. Without again entering upon the details of either case, we beg to note the following quotations from the paper on Pseudo-hypertrophy. "I am convinced that in my patient the disease is essentially a chronic inflammation invading both the perimysium and the muscle fibre." "I may here say that I cannot agree with Gowers and Buss, that the proliferation of connective tissue is the primary, and the disease of the muscular tissue is the secondary process. Either the reverse of this is true or the process occurs simultaneously both in the muscle and the perimysium."[2] These citations sufficiently show the similarity of the findings in the two cases. From the arguments here adduced we can come to no other conclusion than that in this case of polymyositis progression we are dealing with a form of primary myopathy, closely allied if not identical with some forms of primary progressive muscular atrophy.

The following paper was read:

PROGRESSIVE MUSCULAR DYSTROPHIES: THE RELATION OF THE PRIMARY FORMS TO ONE ANOTHER AND TO TYPICAL PROGRESSIVE MUSCULAR ATROPHY.[3]

BY B. SACHS, M D.

The diseases to be discussed in this article have passed under so many different names that it will be necessary, first of all, to state what shall, and what shall not, be understood by the term "Progressive Muscular Dystrophies."

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[1] *Friedreich*, Loc. cit., case 10, p. 37.

[2] *Jacoby, G. W.*, Microscopical studies in a case of pseudo-hypertrophic paralysis, Journal of Nervous and Mental Disease, Vol. xiv., 1887, Sept., Oct.

[3] This article, with full references, was published in the *N. Y. Medical Journal*, December 8 and 15, 1888.